Hemodynamics and systolic and diastolic function in single ventricle

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Single ventricle or univentricular heart

- Hearts for which division into two separate ventricles is deemed surgically infeasible

<table>
<thead>
<tr>
<th>Clinical presentation</th>
<th>Anatomic example</th>
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<tbody>
<tr>
<td>1. Inadequate pulmonary circulation (or ductal-dependent)</td>
<td>- {S,D,S} Tricuspid atresia with pulmonary stenosis (IB)</td>
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<td>- Single right or left ventricle with pulmonary atresia</td>
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<td>2. Excessive pulmonary circulation without systemic outflow obstruction</td>
<td>- {S,D,D} Tricuspid atresia, with transposition of the great arteries and unrestricted pulmonary blood flow (IIC)</td>
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<td>3. Excessive pulmonary circulation with systemic outflow obstruction</td>
<td>- Hypoplastic left heart syndrome (MS/AS; MA/AS; MS/AA; MA/AA)</td>
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<td></td>
<td>- {S,D,D} tricuspid atresia with restrictive VSD</td>
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<td>- {S,L,L} Double outlet left ventricle with restrictive VSD</td>
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<tr>
<td>4. Balanced pulmonary blood flow</td>
<td>- {S,D,S} Tricuspid atresia with pulmonary stenosis (IB)</td>
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<td></td>
<td>- {S,D,D} DORV uncommitted VSD with pulmonary stenosis</td>
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Goals of early managements

• Relief of outflow obstruction
• Optimized Qp/Qs
• Unimpeded pulmonary venous return
• Provision of adequate intracardiac mixing

- Arch reconstruction, aortopulmonary anastomosis, correction of APVR, atrial septectomy, aortopulmonary shunt, PA banding
Circuits in parallel and in series

ventricle

Rs
Rp

RV

LV

Rs
Rp
Arterial saturation and Qp/Qs in single ventricle

![Graph showing arterial saturation (%) against Qp/Qs ratio (0-5)](image)

- Aortic saturations as plotted against Qp/Qs

_Cardiol Young 2003;13:316-22_
Arterial saturation and Qp/Qs in single ventricle

- $\text{SaO}_2$ depends on Qp/Qs
- To achieve $\text{SaO}_2$ 90%, Qp/Qs at least $> 3:1$
- Qp $\uparrow$ to very high level $\rightarrow$ little gain in $\text{SaO}_2$
- At Qp/Qs 1:1$\sim$2:1, Qp $\uparrow$ $\rightarrow$ significant $\text{SaO}_2$ $\uparrow$
### Arterial saturation and Qp/Qs in single ventricle

- SaO$_2$ depends on Qp
- Qp related to Rs

<table>
<thead>
<tr>
<th>To increase Rp</th>
<th>To decrease Rp</th>
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<tbody>
<tr>
<td>Acidosis</td>
<td>Alkalosis</td>
</tr>
<tr>
<td>Inc PEEP</td>
<td>Nitric oxide</td>
</tr>
<tr>
<td>Vasopressor agents</td>
<td>Low MAP</td>
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<tr>
<td>Noradrenaline, adrenaline</td>
<td>High FiO$_2$</td>
</tr>
<tr>
<td>High PaCO$_2$, hypoxia, added nitrogen</td>
<td>Phosphodiesterase inhibitors, nitrates</td>
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</table>
Potential natural history of HLHS

- Ideal anatomy and physiology – Qp/Qs around 1; mild RV volume overload; SaO₂ around 80%; normal Qs
- Falling Rp – Qp/Qs ↑; sign of failure; SaO₂ around 85%
- Low Rp – Qp ↑↑; obvious heart failure with poor peripheral perfusion, metabolic acidosis but high SaO₂
- Closing arterial duct – very low Qs; shock with renal failure; high SaO₂
Systolic functions in palliated univentricular heart

- Result of palliation – volume overload
- $\text{SaO}_2 > 85\% \iff \text{Qp/Qs} > 1.5:1 \iff \text{total ventricular output 2-3 times normal} \iff \text{end-diastolic volume 200-300\% normal}$
- Chronic volume overload – progressive ventricular dilation, more spherical ventricular shape
Systolic function after SCPA

- Persistent cyanosis with mean SaO₂ 75%
- For adequate systemic O₂ delivery, CO↑↑
- End-diastolic volume↓ but still 125-200% normal
- Volume-load reduction, but not normalization
Systolic function after SCPA

- Age-related phenomenon
  - SCPA at 3 yrs → ventricular volume ↓
  - SCPA at 10 yrs → no change

_JACC 1996;28:1301-7_

- Morphologic single LV vs RV
- Prior aortopulmonary shunt vs PA band
Systolic function after TCPA

- Nearly total separation of systemic and pulmonary circulation
- $\text{SaO}_2$ 90-95% or less, ↓ during exercise
- Prior to SCPA or Fontan, $R_s$ 20% below normal ← vasorelaxation due to arterial desaturation and parallel relationship of two circulation
Systolic function after TCPA

- Acute rise in Rs
- Moving the pulmonary circulation into series $\rightarrow$ cross sectional area $\downarrow$, total vascular length $\uparrow \rightarrow$ Rs $\uparrow$
- Peripheral vasoconstriction and neurohumoral activation
- Net impact, 75% increase in Rs
Systolic function after TCPA

- After Fontan, dramatic fall in preload (35-50%, previous SCPA 15%)
- Afterload ↑ and preload ↓ ⇒ systolic function ↓
- Under the optimal circumstances, subsequent ventricular remodeling and somatic growth lead to normalization of ventricular size
Systolic function after TCPA

- Ventricular remodeling with improved ventricular contractility in Fontan at 10 yrs of age, no such recovery in Fontan at a later age

*Circulation* 1992;86:1753-61

- Other factors related to late outcome
  - Prolonged elevation of coronary sinus pressure
  - Valve regurgitation
  - Durability of systemic right ventricle
Diastolic function in palliated univentricular heart

- In tricuspid atresia
- Significant ventricular dilation, reduced mass/volume ratio, normal chamber stiffness

*Am J Cardiol* 1994;73:292-7
Diastolic function after SCPA or Fontan

- Preload ↓ → end-diastolic volume ↓ → mass:volume ratio ↑
- Ventricular compliance ?
- Prolonged isovolumetric relaxation time
- Reduced early rapid filling
Ventricular compliance

- Chronic volume overload → rightward shift of diastolic pressure-volume relationship
- Increased chamber distensibility due to ventricular remodeling
- Chamber compliance is not impaired by acute fall in preload
Impact of ventricular hypertrophy

- *Circulation* 1987;76(Suppl):III45-52
  Reduced mass:volume ratio asso with reduced survival after TCPA

- *J Thorac Cardiovasc Surg* 1986;92:1049-64
  Hypertrophy – longterm risk factor for death after Fontan

- *Herz* 1992;17:220-7
  Lower volume and higher mass:volume ratio in group with poor outcome
Median Age at “Hemi-Fontan”
(Boston Children’s Hospital)

Median Age at Fontan Operation
(Boston Children’s Hospital)
Ventricular relaxation

- Prolongation of isovolumetric relaxation time and lower E:A ratio after Fontan indicating impaired relaxation
- Incoordinate relaxation, inter- and intra-ventricular conduction defect, elevated coronary sinus pressure
Summary

• Palliated single ventricle is at risk for chronic volume overload induced myocardial injury, the severity of which is related to the duration and magnitude of volume overload.

• SCPA and Fontan procedures reduce the volume load on the ventricle, and performed early serve to protect the myocardium from irreversible injury.